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NIH Fiche to Paper Journal

TITLE:	JOURNAL OF CLINICAL NEURO-OPHTHALMOLOGY
PUBLISHER/PLACE:	Raven Press New York Ny
VOLUME/ISSUE/PAGES:	1986 Jun;6(2):109-12 109-12
DATE:	1986
AUTHOR OF ARTICLE:	Barontini F; Maurri S; Sita D
TITLE OF ARTICLE:	Peripheral ophthalmoplegia as the only sign of lat
ISSN:	0272-846X
OTHER NOS/LETTERS:	Library does NOT report holding title 8109051 2942566
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Journal of Clinical Neuro-ophthalmology 6(2): 109–112, 1986.

Peripheral Ophthalmoplegia as the Only Sign of Late-Onset Fibrous Dysplasia of the Skull

F. Barontini, S. Maurri, and D. Sità

A 49-year-old woman presented with a left eye abduction-elevation defect and a bilateral internal rectus palsy of peripheral origin. Thyroid function, cranial computed tomographic scan, and cerebrospinal fluid examination were normal, as were the prostigmine test for myasthenia gravis and the guanidine hydrochloride test for myasthenic syndrome. Skull radiography showed osteosclerotic and osteolytic areas, with slight orbital distortion, and there was hyperactivity in an isotopic scan. A skill biopsy showed fibrous dysplasia. This case is an unusual example of fibrous dysplasia of the skull with neuro-ophthalmological symptoms but without ptosis, exophthalmos, or visual loss.

Diplopia in a patient over 40 years of age often presents diagnostic difficulties, particularly when the more common causes, thyroid myopathy and myasthenia gravis, have been excluded.

In the case described, the diplopia was due to a slight deformation of the orbital cavity caused by the fibrous dysplasia, which had previously been asymptomatic. This rare condition, which gave some diagnostic difficulty because of the absence of exophthalmos and visual defect, should be considered a further cause of "orbital pseudotumor" (1).

CASE REPORT

A 49-year-old woman was admitted on April 2, 1984, with a 3-month history of first transient and then persistent double vision. Two years before, on July 27, 1982, she had had a slight head injury.

The general examination was unremarkable. Abnormalities on the neurological examination were confined to oculomotor function, with a defect in abduction and elevation of the left eye and a slight bilateral internal rectus palsy (Fig. 1).

A chest x-ray film, EKG, electroencephalogram, blood and urine examination, blood sugar curve, and estimation of thyroid function were all normal. A skull x-ray examination 2 years before had shown slight osteosclerotic changes in the left parietal bone. We repeated this test and it showed increased osteosclerotic changes in the fronto-orbital regions and fronto-ethmoidal sinuses and also osteolytic areas in the frontobasal and parietal regions (Fig. 2). A skull computed tomographic (CT) scan confirmed these findings (Fig. 3). An intracranial CT scan with contrast enhancement gave a normal result, and a total body gallium citrate isotopic bone scan showed increased activity in discrete areas of the skull (Fig. 4). The *Treponema pallidum* hemoagglutination test was negative.

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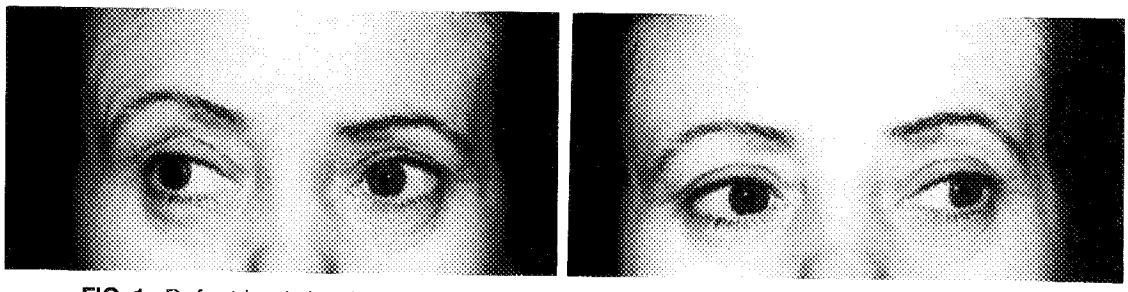

FIG. 1. Defect in abduction and elevation of left eye and slight bilateral internal rectus palsy.

The Lancaster red-green test and a synoptometric examination showed a weakness of the left inferior oblique muscle together with a bilateral internal rectus palsy on attempted horizontal gaze in either direction, suggesting a bilateral internuclear ophthalmoplegia. Pupil diameters were 4 mm on the right and 3 mm on the left, but reactions to light and accommodation were normal. Visual acuity was 20/20 in both eyes. The optic fundi, visual fields, color vision, and light intensity examinations were all normal.

Lumbar puncture revealed a clear cerebrospinal fluid (CSF) under normal pressure, cell count of 5/mm^3, protein level of 63 mg/dl, glucose level of 59 mg/dl, and chloride level of 123 mg/dl. Electrophoresis of the CSF showed no oligoclonal bands in the gammaglobulin fraction and Link's index was normal. Visual-evoked potentials showed normal findings in both eyes.

A test with prostigmine (1 mg i.m.) as well as 10 mg tensilon i.v. was negative. Dexamethasone, 8 mg daily for a week, did not alter the ophthalmoplegia. The otoneurological examination showed absence of spontaneous evoked vestibular signs and the visuo- and vestibulo-oculomotor tests gave normal results.

Serum protein, alkaline phosphatase, acid phosphatase, and calcium levels were all normal. Bence Jones protein was absent in the urine and 24-h urinary calcium and phosphate excretion was normal. Multiple myeloma was thus excluded.

Radiographic and echographic examinations revealed no evidence of neoplastic disease. Guanidine hydrochloride, 500 mg b.i.d., for a week had no effect.

A skull biopsy was performed with the permission of the patient after a few months. Histological examination of the specimen from the right parietal bone, where the isotopic scan had shown increased activity, revealed cellular fibrous tissue with irregular bone formation and infrequent os-

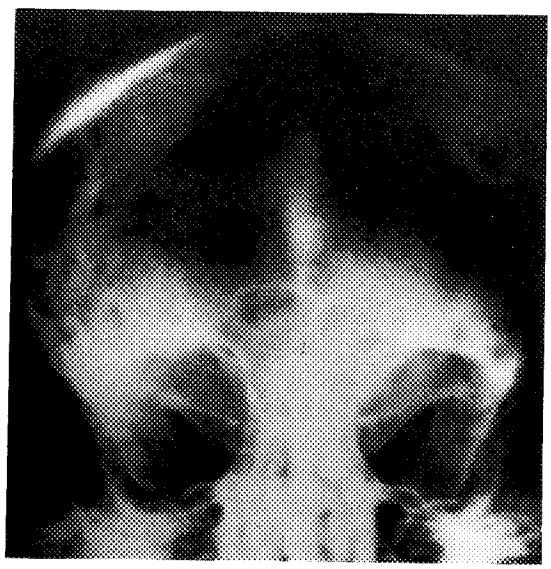

FIG. 2. Skull radiograph showing osteosclerotic changes in the fronto-orbital regions and fronto-ethmoidal sinuses, as well as osteolytic areas in the frontobasal and parietal regions.

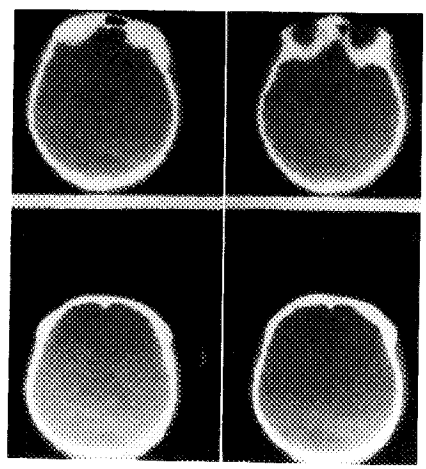

FIG. 3. Computed tomographic scan of skull confirms findings described in Fig. 2.

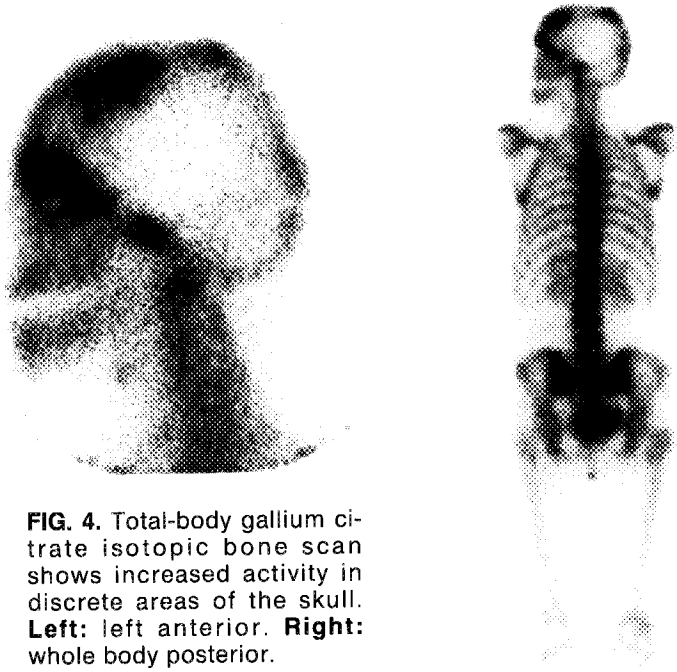

FIG. 4. Total-body gallium citrate isotopic bone scan shows increased activity in discrete areas of the skull. **Left:** left anterior. **Right:** whole body posterior.

teoblastic rimming of the bony trabeculae—the characteristic findings of fibrous dysplasia (Fig. 5).

The patient still complains of diplopia and shows a complex peripheral ophthalmoplegia, with alternating prevalence of the bilateral adduction defect and the abduction-elevation defect of the left eye. Exophthalmos is still absent, both in the CT scan and on examination with the Hertel exophthalmometer (right eye, 22 mm; left eye, 24 mm).

DISCUSSION

In the case we describe, radiography of the skull showed multiple osteolytic areas, the pathological nature of which had to be explained. The appropriate investigations had excluded myelomatosis and metastases, and the possibilities of localized Paget's disease, eosinophilic granuloma (2), and sarcoidosis (3) were considered. However, biopsy of the bone from an area where scintigraphy had

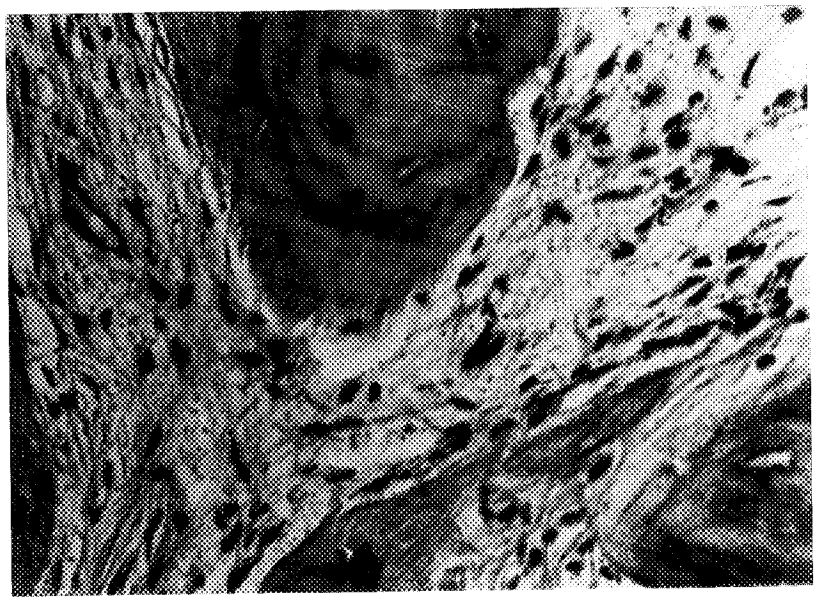

FIG. 5. Histologic specimen from right parietal bone shows cellular fribrous tissue with irregular bone formation and infrequent osteoblastic rimming of the bony trabeculae—characteristic findings of fibrous dysplasia.

shown increased metabolic activity revealed fibrous dysplasia. It is well known that fibrous dysplasia of the skull, or focal areas of "fibrous osteodystrophy," shows histologically a proliferation of intraosseous cellular fibrous tissue (4). In spite of the fact that the condition usually affects patients under the age of 30 years, it has been found in patients over the age of 60 (4).

The fibrous dysplasia may be disseminated or localized, and multiple foci in the skull and in the fronto-orbital regions, as in our case, are not unusual. The most common signs are progressive cranial enlargement and deformity, with distortion of the orbits and exophthalmos. Visual impairment is also common, because of compression of the optic nerve or ophthalmic artery (4,5). Cases have also been described that began with unilateral cranial nerve compression (6) or with involvement of the orbital roof (7) associated with ptosis and loss of visual acuity (6–8).

In our patient, the first sign was a weakness of adduction, but without papilledema, visual loss, ptosis, or exophthalmos. These signs can be explained by the downward and lateral displacement of the eyeballs by the dysplasia, which, although not excessive, involved the inner side of the left orbit and the fronto-orbital region bilaterally enough to deform the orbital contours.

The prognosis of fibrous dysplasia is usually good, but rarely (0.4%) it may undergo malignant transformation (4,7). Progression is said to be unusual after 20–30 years (4). In some cases mentioned in the literature, fibrous dysplasia followed a head injury, but we are doubtful of this cause in our patient.

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